Transcatheter perforation followed by pulmonary valvuloplasty in neonates with pulmonary atresia and ventricular septal defect

Intérêt du cathétérisme avec perforation et valvuloplastie pulmonaire chez les nouveau-nés atteints d’atrésie pulmonaire avec communication interventriculaire

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Summary

Background. — The classic management of neonates with pulmonary atresia with ventricular septal defect (PAVSD) and moderately hypoplastic pulmonary arteries is usually a systemic to pulmonary artery shunt or ductus arteriosus stenting. We report our experience of transcatheter treatment of PAVSD by perforation followed by balloon dilation of the valve, as it is performed in pulmonary atresia – intact ventricular septum.

Abbreviations: PAVSD, pulmonary atresia with ventricular septal defect; PAIVS, pulmonary atresia with intact ventricular septum.

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Patients and method. — Three patients were treated at a mean age and weight of 7.5 days (range 7—8) and 2.9 kg (range 2.5—3.3), respectively. Two newborns were prenatally diagnosed, with micro deletion 22q11 in one case. The three patients had no other pulmonary blood flow support than the ductus arteriosus and were on prostaglandin E1 infusion. The pulmonary atresia was predominantly valvular without significant muscular obstruction. By echocardiography, the mean size of the pulmonary annulus was 6.5 mm (range 6—7). In all cases, the valvular perforation was performed with a 0.014 coronary guidewire, followed by balloon dilation when successfully.

Results. — The procedure succeeded in two cases but failed in the third newborn in whom a long subvalvar muscular stenosis was found at surgery. In the two successful cases, the mean preprocedural transpulmonary doppler gradient was 33.5 mmHg (range 17—50). One patient experienced a femoral venous thrombosis that was successfully treated by heparin and a transient right bundle branch block occurred in another one. In the two successful cases, the prostaglandin E1 infusion was weaned and the surgical repair was performed at 4 and 12 months, respectively.

Conclusion. — In selected cases with PAVSD, perforation followed by balloon dilation of the pulmonary valve offers an interesting alternative to other surgical or transcatheter palliative therapies.

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MOTS CLÉS
Cathétérisme interventionnel ; Atrésie pulmonaire avec communication interventriculaire ; Nouveau-né ; Traitement palliatif

Résumé
Introduction. — Le traitement de l’atrésie pulmonaire à septum interventriculaire ouvert (APVSD) consiste habituellement en une chirurgie néonatale palliative à type de shunt systémico-pulmonaire ou parfois de la mise en place par cathétérisme d’une endoprothèse dans le canal artériel. La correction complète est réalisée après quelques mois. Nous rapportons notre expérience chez ces patients du cathétérisme néonatal avec perforation puis valvuloplastie pulmonaire telle qu’elle est pratiquée dans l’atrésie pulmonaire à septum intact.

Patients et méthode. — Notre recueil rétrospectif depuis janvier 2001 a inclus trois patients d’âge et de poids moyens de 7,5 jours (7—8) et 2,9 kg (2,5—3,3), respectivement. Tous avaient une perfusion de prostaglandines E1, sans autre source de flux pulmonaire que le canal artériel. L’atrésie était principalement valvulaire sans composante musculaire significative associée avec un anneau pulmonaire mesurant en moyenne 6,5 mm (6—7). Les perforations valvulaires ont été effectuées au guide, suivies d’une valvuloplastie au ballonnet.

Résultats. — Le gradient transvalvulaire en fin de procédure était de 33,5 mmHg (17—50) chez deux des patients, la procédure ayant échoué pour le troisième. Lors de sa chirurgie, une longue sténose musculaire a été retrouvée. Aucune complication n’est survenue en dehors d’une thrombose veineuse fémorale résolutive et d’un bloc de branche droit transitoire. La valvuloplastie pulmonaire a permis l’arrêt des prostaglandines E1. La correction chirurgicale a été réalisée à quatre et 12 mois, respectivement.

Conclusion. — La perforation-dilatation par cathétérisme est une alternative intéressante aux autres traitements néonataux dans certaines APVSD. Une plus grande expérience est indispensable pour confirmer l’efficacité et l’innocuité de cette technique.

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Introduction
Pulmonary atresia with ventricular septal defect (PAVSD) comprises a broad anatomical spectrum as evidenced by its classification in four stages [1]. Therefore, there are several possible more or less complex management approaches depending on the significance of lesions [1—4]. The most common approach is the surgical systemic to pulmonary artery shunt during the neonatal period, followed by surgical repair when patients are a few months older [1,5—8]. In a recent study, few cases of PAVSD with valve atresia only, a moderately hypoplastic pulmonary tree, confluent pulmonary arteries and no aortopulmonary collaterals, have been treated with perforation followed by valvuloplasty [9]. This technique is similar to that used in the treatment of pulmonary atresia with intact ventricular septum (PAIVS) [10,11] and offers an interesting alternative to the palliative surgical shunt. Nevertheless, the few cases reported in a single centre are not enough to demonstrate the feasibility of this technique [9], insofar as only the successful cases are described. Moreover, indications for this type of catheterization remain poorly defined.

We report our experience with such management in three newborns with PAVSD.
Patients and method

Patients

From January 2001 to June 2007, three newborns with PAVSD were treated with perforation followed by pulmonary valvuloplasty. The mean age was 7.5 days (range 7–8) and the mean weight was 2.9 kilograms (range 2.5–3.3). Pulmonary atresia was purely valvular with a patent infundibulum and confluent but moderately hypoplastic pulmonary arteries. There were no aortopulmonary collateral arteries. The ductus arteriosus was the only source of pulmonary blood flow support in all three cases, maintained with an infusion of prostaglandin E1.

The mean size of the pulmonary annulus was measured at 6.5 mm (range 6–7), while the pulmonary trunk was 6.4 mm (range 6–6.8), the right pulmonary artery was 3.75 mm (range 3.5–4) and the left pulmonary artery was 3.8 mm (range 3.6–4) on the echocardiography.

One of the patients was a carrier of microdeletion 22q11 while the other two had been prenatally diagnosed.

Interventional catheterization

Under general anaesthesia and through femoral vein access, the initial step consisted in a right ventriculogram on lateral view, using a right Judkins 2 or 2.5 curve end-hole coronary catheter (Judkins, Cordis Corporation, NJ, USA). This angiography performed immediately under the valve annulus confirmed the pulmonary atresia and its purely membranous origin (Fig. 1). The 4F right coronary catheter (Judkins, Cordis Corporation, NJ, USA) is positioned just below the valve membranous atresia. Two guidewires (BMW 0.014 Boston Scientific, Natick, MA, USA) are concomitantly inserted in the 4F right coronary catheter, one through its rigid tip and the other one through its soft tip. A contrast medium injection through a ‘‘Y’’ connection enables to control the perpendicular positioning of the guidewires in relation to the valve atresia (Fig. 2). The perforation is then performed with the rigid end of the guidewire, which is advanced 0.5 cm to 1 cm through the valve annulus (Fig. 3). The flexible-end guidewire is then positioned in the pul-
Figure 4. Lateral view angiogram in the infundibulum: visualization of the guidewire in the pulmonary arterial tree after perforation of the valve.

Figure 5. Lateral view showing pulmonary valvuloplasty with a 6 mm × 20 mm balloon.

In the other one. Infusion with PGE1 could be discontinued in the two successful cases with closure of the arterial duct a few days after catheterization.

Biventricular surgical repair was successfully performed at 4 months for one patient and 12 for the other. Preoperative catheterization objectified good development of the pulmonary arteries (Fig. 6).

After a follow-up of 13 and 21 months, neither of these two patients needed any additional surgery or interventional catheterization. The mean hospital length of stay for these children during surgical repair was 10.5 days (range 10–11).

Results

In one case, perforation of the pulmonary atresia failed due to the narrowness and length of the infundibulum which did not allow correct positioning of the wire. A Blalock-Taussig surgical anastomosis was performed in this patient a few days later. During corrective surgery at 8 months, the infundibulum was described as hypertrophied and particularly narrow.

For the other two cases, the procedure was successful. The main results are reported in Table 1.

One patient experienced a femoral vein thrombosis, which resolved with anticoagulation, and a transitory right bundle branch block was observed following catheterization in the other one. Infusion with PGE1 could be discontinued in the two successful cases with closure of the arterial duct a few days after catheterization.

Biventricular surgical repair was successfully performed at 4 months for one patient and 12 for the other. Preoperative catheterization objectified good development of the pulmonary arteries (Fig. 6).

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Discussion

Transcatheter perforation of the pulmonary valve using radiofrequency or with a guidewire had already been widely described for PAIVS in the neonatal period [9–11]. The first case was reported in 1991 by Qureshi et al. [10]. Since then, several retrospective studies confirmed its feasibility in 80 to 90% of cases with failures due to the impossibility of perforating the valve [10,11]. Our results confirmed its indication and feasibility for PAVSD where atresia is purely valvular and associated with hypoplasia of the pulmonary arterial tree. Hence, perforation followed by balloon dilation do

Table 1 Characteristics of the two successful pulmonary valve perforation.

<table>
<thead>
<tr>
<th>Pts</th>
<th>Balloon (diam)/guidewire</th>
<th>Gdt</th>
<th>KT length (min)</th>
<th>Complications</th>
<th>SaO2 (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Tyshak(^a) 6 mm/0.014 BMW(^b)</td>
<td>50</td>
<td>123</td>
<td>Transient RBBB</td>
<td>85</td>
</tr>
<tr>
<td>2</td>
<td>Tyshak(^a) 6 mm/0.014 BMW(^b)</td>
<td>17</td>
<td>92</td>
<td>Fem V thrombosis</td>
<td>90</td>
</tr>
</tbody>
</table>

Pts: patients; RBBB: right bundle branch block; diam: diameter; Fem: femoral; Gdt: postcatheterization peak-to-peak gradient; KT: catheterization; min: minutes; SaO2: transcutaneous oxygen saturation after catheterization and closure of arterial duct; V: vein.

\(^a\) Tyshak, NuMed, NY, USA.

\(^b\) BMW, Boston Scientific, Natick, MA, USA.
Transcatheter perforation and valvuloplasty in pulmonary atresia - ventricular septal defect

Therefore, the perforation and balloon dilation technique appears to be contraindicated. Only few reports have been published [2,12—15], with the largest study including eight cases over a period of six years [14]. In the case of associated infundibulum narrowing, the procedure was associated with stent implantation [2,13,14]. Our study suggests that this technique may only apply to PAIVSD with predominantly valvular atresia, confluent but moderately hypoplastic arteries and ductus arteriosus dependant pulmonary blood flow with no further supply from the aortopulmonary collateral arteries.

The diversity in the management of PAIVSD reflects the heterogeneous nature of the lesions [3,4]. When the pulmonary arteries are profoundly hypoplastic with an aortopulmonary collateral dependant pulmonary blood flow, various palliative and curative surgical interventions such as one stage unifocalization of the pulmonary arterial tree are performed [4]. When the pulmonary arterial tree is well developed and dependent on the ductus arteriosus thereby requiring infusion of prostaglandins E1, there are several therapeutic options. Rarely, surgical correction is performed in the newborn [16]. In the majority of the centres, an initial palliative step is performed in the neonate followed by corrective surgery 4 to 12 months later. This palliation may be surgical with a modified Blalock-Taussig anastomosis, which may be complicated by distortion of the pulmonary arteries, particularly if they are hypoplastic [3,5,6,17]. It may also include interventional catheterization with stent implantation in the arterial duct. Nevertheless, the latter is often technically difficult in PAIVSD that is readily associated with a tortuous arterial duct [18]. In addition, the pulmonary blood flow is provided in an asymmetrical and partially retrograde direction, that is less favourable to pulmonary artery growth than an antegrade flow [19].

Figure 6. Frontal pulmonary angiography at 12 months of age showing good development of the pulmonary arteries before surgical repair.

Historically, this perforation followed by balloon dilation strategy is inspired by dilation of the pulmonary valve in the tetralogy of Fallot suggested about twenty years ago. While this remains a controversial procedure, encouraging results have been published. Yet, indications for such procedure remain limited as this technique concerned the rare patients with a predominantly valvular obstruction. Moreover, the development of successful surgical repair for tetralogy of Fallot earlier in life further limit the indications for balloon dilation of the pulmonary valve [20]. Similarly to balloon dilation, perforation followed by pulmonary valvuloplasty is an interesting alternative in rare cases of PAIVSD with moderately hypoplastic but confluent pulmonary arteries, since an antegrade and pulsatile flow offers a better growth potential for the pulmonary arteries [2,12—14]. The induced pulmonary insufficiency also contributes to their development. In addition, this treatment enables an easier access to the pulmonary arterial tree if further diagnostic or interventional catheterization is indicated, for example, in the case of branch pulmonary stenosis [14]. Finally, surgical repair is performed at an age when its morbidity and mortality are lower [2,12—14]. The assessment and extensive use of this technique are restricted by the low number of patients concerned. Moreover, the experience reported on the perforation of PAIVS revealed that complications occur in approximately 15% of cases, while the most common and dangerous include cardiac perforation responsible for rare deaths [10,11]. During the perforation procedure, the wire may perforate the infundibulum, which most often does not lead to major pericardial effusion [13,14]. However, in the Canadian study, the percentage of right ventricular perforation was close to 50% with haemodynamic instability in one patient [14]. In this setting, the use of biplane system enabling to visualise the perforation through frontal and lateral plan is mandatory. In addition, the use of radiofrequency instead of the stiff tip of a guidewire could decrease the rate of perforation. Finally, vascular complications including femoral thrombosis, blood loss with the need blood transfusions are potentially more common in newborns. All this reinforces the concept that this type of interventional catheterization should only be practised by very experienced teams.

Conclusion

Interventional catheterization with perforation and dilation of the pulmonary valve offers an interesting alternative to other palliative therapies for some cases of PAIVSD with moderately hypoplastic but confluent pulmonary arteries and a patent infundibulum. It only applies to a small number
of patients. Its indication should be discussed according to individual cases and in relation to the benefit/risk ratio compared with surgery, which remains the reference method. In all cases, it must be performed by an experienced team.

**Conflict of interests**

There is no potential conflict of interest.

**References**


